# Triple Positive Anti-phospholipid syndrome: A curious case of cutaneous necrosis

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## Learning Objectives

- Recognize cases of Antiphospholipid syndrome (APS) without common risk factors.
- Diagnose and treat cutaneous necrosis in APS.

## Case Presentation

- 50-year-old female with hypothyroidism, and chronic venous insufficiency presented with altered mental status
- Exam was significant for lethargy, bilateral lower extremity edema, and necrotic weeping wounds with purulent discharge.
- She was admitted for toxic metabolic encephalopathy secondary to severe sepsis, evident by leukocytosis (26.47K/UL), lactic acidosis (3.5MMOL/L), hypotension (68/47mmHG), and tachycardia (110bpm).
- The infection, initially presumed to be cellulitis related to bilateral lower extremity wounds, prompted initiation of empirical antibiotic therapy. Subsequent deterioration required transfer to the ICU for hemodynamic support.
- Despite improvement in mental status with antibiotics, the lower extremity wounds continued to deteriorate, expanding to her upper thighs with worsening necrosis.
- She became thrombocytopenic (nadir of 52 K/UL) which prompted further testing. DIC workup was negative, CT scan of the lower extremities was normal and no deep vein thrombosis (DVT) was seen on doppler. Blood cultures remained negative.

## Image

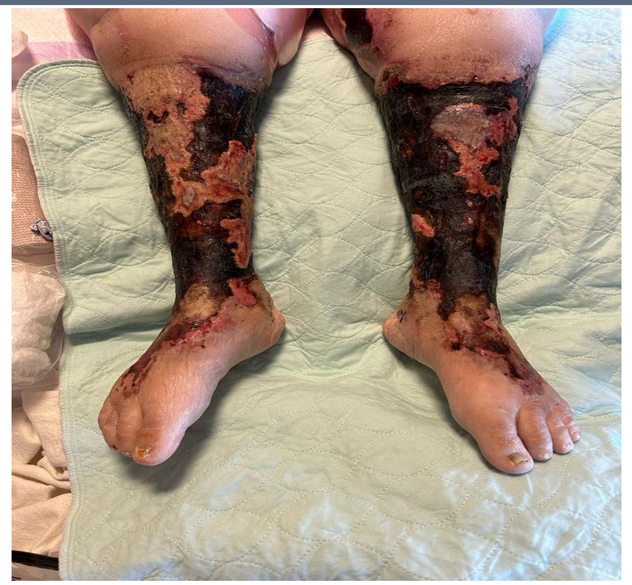

***Figure 1*** Bilateral lower extremities with *extensive cutaneous necrosis* seen

## Case continued

- She eventually tested positive for beta-2 glycoprotein, anti-cardiolipin antibodies, and lupus anticoagulant. Skin punch biopsy of the lesions confirmed cutaneous infarction with underlying vascular thrombosis.
- This, along with APS antibodies clinched our diagnosis.
- Treatment with high-dose steroids and therapeutic anticoagulation was initiated. Consequently, the patient was considered stable for discharge with ongoing wound care and potential surgical intervention.
- Confirmation of her APS diagnosis will be done at the 12-week mark.

## Discussion

- APS is a rare autoimmune disease with an annual incidence of 1 to 2 per 100,000.
- It presents with a wide array of cutaneous findings such as livedo reticularis and ulcers, with disseminated cutaneous necrosis being the rarest.
- It is not considered in common differentials when patients present with diffuse cutaneous findings
- In our case, it was even harder to diagnose since our patient *lacked* the common risk factors of multiple pregnancy losses and previous history of DVT.
- Cutaneous findings must be addressed early on since there is a significant morbidity rate
- 60% need surgery for debridement or amputation] and highly associated with catastrophic APS (CAPS), with risk of progression to other vital organs.
- There is no established criteria for managing cutaneous necrosis in APS
- Our patient responded to therapeutic anticoagulation and high-dose steroids with surgical debridement scheduled.

## References

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